Retroperitoneal extra-adrenal myelolipoma: Appearance in CT and MRI

N. Butori\textsuperscript{a,\textdagger}, F. Guy\textsuperscript{a}, F. Collin\textsuperscript{b}, C. Benet\textsuperscript{b}, S. Causeret\textsuperscript{c}, N. Isambert\textsuperscript{d}

\textsuperscript{a} Department of Medical Imaging, Georges-François-Leclerc Center, 1, rue Pr-Marion, BP 77980, 21079 Dijon cedex, France
\textsuperscript{b} Laboratory of Pathological Anatomy and Cytology, Georges-François-Leclerc Center, 1, rue Pr-Marion, BP 77980, 21079 Dijon cedex, France
\textsuperscript{c} Department of Surgery, Georges-François-Leclerc Center, 1, rue Pr-Marion, BP 77980, 21079 Dijon cedex, France
\textsuperscript{d} Department of Medical Oncology, Georges-François-Leclerc Center, 1, rue Pr-Marion, BP 77980, 21079 Dijon cedex, France

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A myelolipoma is a rare benign lesion often discovered by chance during a tomodensitometry (CT) examination of the abdomen. It is classically located in the adrenal glands and is easily recognised due to its contingent of adipocytes [1]. Extra-adrenal forms are unusual and cause diagnostic difficulties even with histopathology. We report here the case of a male patient with a retroperitoneal extra-adrenal myelolipoma, which presented very much like a well-differentiated liposarcoma. This observation is the moment to recall the imaging characteristics of retroperitoneal fatty tumours and to emphasize the major role of identifying cytogenetic and molecular abnormalities in characterising them.

Observation

A 55-year-old man, with no notable medical history apart from mood and behavioural disorders, was admitted to the dermatology unit for management of exanthema of the left leg that had been evolving for several months. He had no somatic symptoms apart from pruritus associated with his skin rash.

Abdominal ultrasonography, performed to look for the cause of the exanthema, revealed the presence of a hyperechoic mass under the right kidney. An abdominal CT examination confirmed that there was a voluminous, well-delineated, right retroperitoneal mass, of 10 cm at its widest point, predominantly of dense fatty material, with a second tissue component in the form of poorly demarcated layers, enhanced following intravenous injection of an iodinated contrast agent (Fig. 1). The kidneys and adrenal glands were

\textsuperscript{\textdagger} Corresponding author.
E-mail address: butori.noemie@gmail.com (N. Butori).

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normal. The diagnosis suggested was of a malignant retroperitoneal tumour, such as a well-differentiated liposarcoma (WLD). Several CT guided percutaneous microbiopsy samples were taken to confirm this.

The histological analysis revealed a lesion formed of mature adipocytes and fibrous septa, altered by oedema and polymorphous inflammatory cells. There was no cellular atypia, and the immunohistochemical study with anti-MDM2 and anti-CDK4 antibodies was negative. Since there were no histological and phenotypic characteristics of liposarcoma, it was hypothesised that the mass was a simple lipoma; however, the quantity of tissue analysed was small and the radiological characteristics leaned more towards a malignant lesion.

Faced with this inconsistency between the anatomical and radiological findings, abdominal MRI was performed. It did not provide any new semeiotic element but confirmed the coexistence of a fatty component with another quantity of tissue forming zones of moderate enhancement, giving weight to the hypothesis of a malignant fatty lesion (Fig. 2).

Since there was no specific diagnosis based on the histology, but with radiological and probabilistic reasons (the high probability of malignancy of a retroperitoneal fatty mass) for considering it to be a liposarcoma, it was decided following a multidisciplinary consultation to undertake surgery for initial management of a well-differentiated retroperitoneal liposarcoma.

R0 enlarged exeresis of the retroperitoneal mass was undertaken with right nephrectomy and monobloc right hemicolectomy. The pathological anatomy analysis of the mass confirmed the presence of a well-defined lesion of 10.5 × 7 × 3.5 cm, not adhering to either the kidney or the right adrenal gland, limited by a connective capsule and composed of both adipocytes and a haemopoietic contingent, which therefore produced the diagnosis of ectopic retroperitoneal myelolipoma (Fig. 3).

**Figure 1.** Abdominopelvic CT scan following injection of iodinated contrast agent. It shows a well-delineated right subrenal retroperitoneal mass, surrounded by a fine capsule. Two components are visible: a predominant one of fat and the other of tissue, infiltrating the fat and with indistinct contours (a and b). Reformation (b) shows the normal right adrenal gland (arrow) and separated from it, the extra-adrenal mass (head of arrow).

**Figure 2.** Abdominal MRI. Within the retroperitoneal mass, the fatty component (black arrow) can be perfectly identified: in T1 hypersignal it looks identical to that of the subcutaneous fat (a), and is cancelled out (b) by the technique of selective saturation of the fat (FatSat). The tissue component (white arrow) is moderately enhanced after injecting contrast agent (b): a: TSE (turbo spin echo) T1-weighted transverse sequence, TR: 550 ms, TE: 11 ms; b: GRE (rapid gradient echo) T1-weighted transverse sequence, TR: 3.2 ms, TE: 1.1 ms, θ: 50°, with selective saturation of the fat signal (FatSat) and after injection of gadolinium (Dotarem®).
Discussion

A myelolipoma is a rare benign tumour, composed of mature adipose tissue associated in variable proportions with normal haemopoietic tissue.

It is classically asymptomatic and found in a normal adrenal gland. It presents no problem of diagnosis for imaging because it is the only adrenal tumour composed of a quantity of adipose tissue which can always be recognised: in CT, the spontaneous density of this tissue is lower than –30 Hounsfield units, and in MRI, its signal is hyperintense in T1 weighting and is cancelled out following selective saturation of the signal from the fat by FatSat (a signal parallel to that of mature fat). Chemical shift sequences do not objectify any reduction in the opposed-phase signal (unlike suprarenal adenomas which are composed of non-adipocyte cells and are more or less rich in intracellular lipids) [2]. It is necessary and adequate, therefore, to detect adipose tissue within an adrenal nodule to be able to diagnose an isolated adrenal myelolipoma, a lesion that does not require any treatment or special monitoring [1].

Diagnosing a retroperitoneal extra-adrenal myelolipoma, which is much rarer, is more difficult. A fatty retroperitoneal mass could essentially be a retroperitoneal liposarcoma, an adrenal myelolipoma, a renal angiomylipoma or a retroperitoneal teratoma. If there is no specific aetiological guidance, samples must be taken for histological analysis.

In our patient, it was not possible from the histological study of the first samples taken percutaneously to diagnose a myelolipoma, since the three haemopoietic lineages were not detected in the sample. At the end of our investigations, we therefore discussed differential diagnoses of lipoma and WDL.

It is necessary to be able to differentiate these two entities because that decides the management. Indeed, WDL, unlike lipoma, has a risk of locoregional recurrence and potential for transforming into a very highly malignant sarcoma. Wide excision enlarged to the adjacent organs is therefore recommended [3].

Imaging contributes to this differentiation: indeed, tumour size greater than 10 cm, the presence of septa of thickness more than 2 mm or a nodular zone, and a fatty component less than 75%, are highly suggestive of malignancy [4]. For Gaskin and Helms, the sensitivity of MRI is 100% in diagnosing WDL, but its specificity is only 83%, because certain simple lipomas or lipoma variations cannot be distinguished from a WLD [5]. Any fatty lesion that does not look like a simple lipoma in imaging (or have the characteristic appearance of certain benign fatty tumours) should be biopsied to clarify what kind it is.

In histology, distinguishing a lipoma from a WLD can be difficult: it relies on the presence, within the WDL, of immature adipose cells (lipoblasts) and abnormal cells, which are sometimes few in number and heterogeneously distributed. For some years, differential diagnosis between lipoma and WDL has been facilitated by cytogenetics and molecular biology [6], because chromosomal and molecular abnormalities have been consistently and specifically identified in liposarcoma cells. These abnormalities are supernumerary, giant chromosomes and ring chromosomes that carry an amplification of the MDM2 gene, causing hyperexpression of the protein, detectable by immunohistochemistry. It is therefore possible to detect a specific molecular abnormality of liposarcomas on a routine basis.

The diagnostic value of the absence in our patient of any amplification of the MDM2 gene was not known. The hypothesis of a WDL was considered the most probable diagnosis, whereas it should have been excluded, with consequences on the extent of surgical ablation: the right colectomy and even the nephrectomy could have been avoided.

Two lessons have been drawn from this observation. Firstly, there are atypical presentations of myelolipoma, in particular occurrence in extra-adrenal locations, the unusual nature of which justifies histological evidence. This is obtained provided that the samples analysed contain the three haemopoietic lines, sometimes well concealed within the adipose tissue.

Secondly, this clinical case illustrates the contribution made by cytogenetics and molecular biology to the diagnosis of fatty tumours. It is indeed possible to confirm or quash the diagnosis of liposarcoma owing to immunohistochemical detection of hyperexpression of the protein MDM2.

Finally, we would like to highlight another advantage of the techniques related to MDM2: they allow a dedifferentiated liposarcoma to be recognised, and thus certain non-differentiated mesenchymal tumours to be connected to the adipocyte line, which, by default, are qualified as malignant fibrous histiocytomas, sarcomas with a much bleaker prognosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References

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